P-109

Left ventricular noncompaction in children and adolescents: clinical features, treatment and follow-up

Dindar A(1), Ergul Y(1), Nisli K(1), Varkal M.A.(1), Oner N(2), Dursun M (1), Aydogan U(1), Omeroglu R.E(1).
Istanbul University, Istanbul Faculty of Medicine, Istanbul, Turkey (1)
Kartal Kosuyolu Cardiovascular Research and Training Hospital, Istanbul, Turkey (2)

Abstract
Objective: Left ventricular noncompaction (LVNC) is a specific cardiomyopathy that occurs following a disruption of endomyocardial morphogenesis. This study presents clinical findings, diagnostic features, treatment and follow-up of pediatric patients diagnosed with LVNC.
Methods: Patients with LVNC who were followed from January 2006 to March 2010 were included in this study. Diagnosis was made with the use of characteristic findings of magnetic resonance imaging and echocardiography. Holter electrocardiography and metabolic screening tests were also performed in all patients.
Results: A total of 24 patients were studied (18 male, 6 female). Patient age at diagnosis was 50±60 months (8 days to 15 years). Average follow-up period was 22±12 months (4 months to 4 years). Findings at diagnosis were as follows: 8 (33%) patients had heart failure, five (20%) had rhythm abnormalities, five (20%) had cardiomegaly, two had murmurs, two – cyanosis, and two presented with fatigue. Ten (41%) patients had been followed previously with other diagnoses. In 21 (87.5%) patients, electrocardiographic abnormalities were noted, especially left ventricular hypertrophy and ST-T changes. Patients had an average ejection fraction of 46% (18-73%) and three of them had additional congenital heart disease (patent ductus arteriosus, aortopulmonary window and complex cyanotic heart disease). Scanning for metabolic diseases revealed fatty acid oxidation disorder in one patient, and mitochondrial disease in another. During follow-up, a permanent pacemaker was implanted in a patient with severe bradycardia and ventricular dysfunction, and 3 patients died.
Conclusion: LVNC can be diagnosed at any age from newborn to adolescent and has a variable clinical course. Closer study of patients with cardiomegaly and heart failure can reduce delays in diagnosis of LVNC.